A New Syndrome of Hereditary Congenital Corneal Opacities, Cornea Guttata, and Corectopia

Bilateral infantile corneal opacities can be caused by anterior segment dysgenesis, corneal dystrophy, congenital glaucoma, inflammation, systemic storage disease, or sclerocornea.1-3 We describe a mother and son with bilateral superior V-shaped corneal opacities, cornea guttata, and corectopia. This unique combination of anterior segment abnormalities has not, to our knowledge, been reported to be inherited.

Report of Cases. Case 1. A 14-day-old term male infant had bilateral congenital corneal opacities. He could not fix or follow a light source. Slitlamp examination showed dense bilateral superior V-shaped corneal opacities (Figure 1). In the area of opacification, there were a few iridocorneal adhesions and what appeared to be cornea guttata in both eyes. The pupil was drawn superiorly and was occluded by the corneal opacities in both eyes. The lens and fundus could not be visualized. There were no signs of inflammation, systemic dysmorphosis, or an intrauterine-derived infection. Preoperative ultrasonography revealed a normal lens and vitreous and no retinal detachment. Inferior iridectomies were performed in both eyes 2 weeks apart starting at the age of 1.5 months. At the latest follow-up examination 2 years later, he had nystagmus but appeared to have reasonably good vision. His refraction was +2.00 +2.00 × 180 OD and +1.50 +1.25 × 90 OS. The intraocular pressure was normal. There was a mild decrease in the density of the corneal opacities but no change in their surface area during the follow-up period. The optic nerve heads appeared to be smaller than average but were not flagrantly hypoplastic.

Case 2. The mother of the patient in case 1, at the age of 31 years had a best-corrected visual acuity of 20/40 OD (refraction, −11.25 +5.00 × 120) and 20/60 OS (refraction, −15.00 +5.25 × 90). She had full-thickness corneal opacities similar in location but less extensive than those of the infant in case 1. At the age of 9 years, full-thickness corneal opacities were noted in her right eye while the left eye remained clear. At the age of 10 years, full-thickness corneal opacities were noted in her left eye.

4. Hawkins AS, YuJ, Hamming NA, Rubenstein JB. Topical interferon τ/H9251 and perilesional injection and topical interferon α2B.2 These investigators also used a topical dose of 1 mIU/mL but only continued therapy until the lesion was not clinically detectable.1,7

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in her son. She had diffuse corneal guttata and a slightly up-drawn pupil in both eyes (Figure 2). Intraocular pressure was normal. Indirect ophthalmoscopy revealed a normal retina in the right eye and several peripheral inferior chorioretinal scars in the left eye. The nature of the chorioretinal scars could not be resolved. She had neurologic signs and symptoms suggestive of multiple sclerosis. There were no other family members with similar ocular abnormalities.

Comment. This family presents a unique combination of corneal findings that do not fit into known diseases or syndromes. There is some overlap with Peters anomaly, which is usually a sporadic condition characterized by central corneal opacity and defects in the corneal endothelium, Descemet membrane, and posterior stroma.1-5 In our 2 patients, the corneal opacities were superior and peripheral, and there were no visible defects in Descemet membrane. Furthermore, the disease trait appeared to be inherited in an autosomal dominant fashion. Axenfeld-Rieger anomaly is also autosomal dominant and includes posterior embryotoxon, iris stromal hypoplasia, or polycoria; neither had glaucoma. Their corneal opacities were localized and did not progress in surface or density, unlike those of corneal dystrophies or storage diseases. Cornea guttata, such as that in our patients, has not been reported with any of the previously mentioned conditions, to our knowledge.

Optical sector iridectomy is a simple and safe procedure that can improve visual outcome and avoid penetrating keratoplasty.4 This procedure was highly effective in our younger patient with corectopia and peripheral corneal opacities.

Jeong-Min Hwang, MD
Daniel C. Chung, DO
Elias I. Traboulsi, MD
Cleveland, Ohio

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Corresponding author and reprints: Elias I. Traboulsi, MD, Center for Genetic Eye Disease, Cole Eye Institute, the Cleveland Clinic Foundation, i32, 9500 Euclid Ave, Cleveland, OH 44195 (e-mail: traboue@ccf.org).


Acute Anterior Uveitis and Corneal Edema Associated With Travoprost

Prostaglandin analogues have dramatically changed many clinicians’ approach to glaucoma treatment. The combination of potency, once-daily dosing, and relatively few side effects make these appealing agents. The most frequently reported adverse reaction with travoprost is ocular hyperemia (occurring with a frequency of 35%-50% in populations studied). Decreased visual acuity, eye discomfort, foreign body sensation, pain, and pruritis are reported to occur with a frequency of 5% to 10%.1 Herein we report a case of acute iritis and corneal edema after only 5 doses of travoprost.

Report of a Case. A 79-year-old white man had a history of atrial fibrillation and bladder carcinoma. The medications he was taking included digoxin, warfarin sodium, and verapamil. In 1983 he underwent planned extracapsular cataract extraction without an implant lens in the left eye. This was followed 1 year later by phacoemulsification with a posterior chamber intraocular lens placed in the right eye and a retinal detachment repair performed in his left eye. He was diagnosed as having open-angle glaucoma in both eyes in 1983. The glaucoma was well controlled medically over nearly 2 decades. Mild corneal guttata were first noted in 1993.

We saw him on January 2, 2002, for an eye examination. The visual acuity was 20/40 OD and 20/230 OS (without aphakic correction). Findings on slitlamp examination revealed clear corneas with 1-2+ guttata OU. The anterior chambers were quiet. The intraocular pressure was 20 mm Hg OD and 23 mm Hg OS while receiving a therapeutic regimen of 0.5% timolol maleate and bri-